NEUROPSYCHOLOGICAL REHABILITATION
2009, 19 (2), 291–314

Psychology Press
Taylor & Francis Group

Topographical disorientation in a patient who never developed navigational skills: The (re)habilitation treatment

Chiara Incoccia[1], Luisa Magnotti[1], Giuseppe Iaria[2], Laura Piccardi[1,3], and Cecilia Guariglia[1,4]

[1]IRCCS, Fondazione Santa Lucia, Roma, Italy; [2]Faculty of Medicine, University of British Columbia, Vancouver, Canada; [3]Facoltà di Psicologia, Università degli Studi dell'Aquila, Italy; [4]Dipartimento di Psicologia, Università di Roma "La Sapienza", Roma, Italy

Although many cases of topographical disorientation are described in the literature, very few attempts have been made to rehabilitate this deficit, most likely because it is a multi-faceted syndrome in which different patients are affected by different topographical deficits. Therefore, it is not easy to develop a single rehabilitation programme to improve all types of topographical disorders. We describe the rehabilitation of a young woman with selective and pervasive topographical disorientation who never developed navigational skills due to a cerebral malformation bilaterally involving the retrorolandic regions. During treatment, the patient was trained to explore her surroundings carefully, to orient herself and then to move in the environment using a language-based strategy. At the end of the treatment, the patient was able to navigate in the environment by adopting several cognitive strategies useful for orientation. This result was maintained at the one-year follow-up, at which time the patient was also able to reach locations she had never been to alone. These results suggest that even patients who have never developed the ability to orient themselves in the

Correspondence should be sent to Chiara Incoccia, IRCCS, Fondazione Santa Lucia, Centro Ricerche Neuropsicologia, Via Ardeatina 306, CAP 00179, Roma, Italy.
E-mail: c.incoccia@hsantalucia.it

This study was partially supported by grants from the Ministero della Salute RC05 and the European Community FPS-Strep-Wayfinding-N.012959. The authors would like to thank Dr. Filippo Bianchini for his help in preparing figures and collecting data and Dr. Laura Zompanti for her help in evaluating the patient.

http://www.psypress.com/neurorehab DOI:10.1080/09602010802188344

environment can still achieve very good functional recovery if they are accurately assessed and submitted to a specific rehabilitation intervention.

Keywords: Human navigation; Topographical disorientation; Rehabilitation of topographical disorders; Congenital cognitive deficits.

INTRODUCTION

In humans, topographical orientation is a complex brain function that relies on both the different strategies individuals use to move in the environment and on familiarity with the features of the environment. Such a complex, multicomponential skill requires the integration of different cognitive abilities and, consequently, involves several neural systems. Therefore, damage to brain regions that affect different cognitive functions involved in orientation can result in several topographical orientation disorders (Aguirre & D'Esposito, 1999) that are very disabling in everyday life. In fact, patients affected by topographical disorientation may be unable to orient themselves in the environment or to recognise, describe or learn pathways even in familiar surroundings. Although many cases of patients affected by topographical disorientation have been described in the literature (for reviews see Aguirre & D'Esposito, 1999; Barrash, 1998; Farrell, 1996), there are very few reports of rehabilitation of this disorder (Brunsdon, Nickels, Coltheart, & Joy, 2007; Davis & Coltheart, 1999). Several factors contribute to the paucity of information in this area. First, topographical disorientation is commonly reported in patients who have lost the ability to orient themselves in the environment as a consequence of a general neurodegenerative syndrome or a confusional state and who are usually sent to more general rehabilitation programmes (e.g., Reality Orientation Therapy, Taulbee, & Folsom, 1968). Second, in focal brain damaged patients topographical disorientation may be accompanied by more general amnesic disorders; in this case, the inability to memorise paths and environments is treated together with the amnesic deficit. Finally, in cases of selective topographical disorientation the nature of the deficit may also vary greatly among patients (see Aguirre & D'Esposito, 1999), making it difficult to develop a single rehabilitation procedure to deal with several underlying impairments. When a specific rehabilitation treatment has to be developed, however, it is essential to understand the various cognitive impairments in orientation and their relationships with different types of brain damage.

In 1982, De Renzi suggested that topographical disorientation should not be considered a unitary disorder because, in most cases, it depends on other cognitive deficits. First, patients may show topographical disorientation due to perceptual and cognitive processes that compromise the ability to scan the environment visually and to shift attention to different visual targets, as occurs in patients affected by the hemineglect syndrome. Second, patients may show a deficit in localising objects and in correctly perceiving specific features, such as depth and shape, which affect their ability to generate and manipulate visual images. Third, patients may show topographical disorientation due to memory problems; in fact, different memory systems may be involved depending on the strategy used to move in the environment and on the different kinds of material used to become oriented in the environment. De Renzi (1982) also underlined that "pure" topographical disorientation may be characterised by the presence of selective amnesic (failure to remember directions) or agnosic (failure to recognise landmarks) deficits.

In a more recent review, Aguirre and D'Esposito (1999) presented a detailed taxonomy of topographical disorientation by identifying four different categories of disorders. Patients affected by *egocentric disorientation*, following lesions of the posterior parietal cortex, are unable to use egocentric coordinates or to localise environmental landmarks; i.e., although they are able to recognise a landmark, they cannot code its position relative to themselves (see patient GW; Stark, Coslett, & Saffran, 1996). Patients with *heading disorientation*, due to lesions of the retrosplenial cortex (posterior cingulate), recognise landmarks in the environment but are unable to derive directional information from them (see Case 2, Takahashi, Kawamura, Shiota, Kasahata, & Hirayama, 1997). *Landmark agnosia*, due to lesions of the medial occipital-temporal cortex (including fusiform, lingual and parahippocampal gyri), is a more selective damage that prevents patients from recognising salient environmental landmarks (see Case AH, Pallis, 1955). Finally, *anterograde disorientation*, associated with lesions of the parahippocampal cortex, is the inability to learn pathways in novel environments (see Case 1, Habib & Sirigu, 1987). This taxonomy seems to account for patients with a pure topographical disorder, that is, with specific deficits that do not allow correct navigation and orientation.

In our opinion, to assess the topographical disorder, both of the above described models need to be considered. Specifically, a general neuropsychological evaluation, together with an assessment of several cognitive strategies useful for orientation, should help in developing a specific rehabilitation treatment for recovering topographical skills by strengthening unaffected strategies in order to bypass defective ones.

To our knowledge there are only two published reports on the treatment of topographical disorientation (Davis & Coltheart, 1999; Brunsdon et al., 2007). Davis and Coltheart (1999) described the treatment of a patient with topographical disorientation who was mainly affected by a memory deficit; the rehabilitation therapy was based on learning the names of streets and their positions on the map and recognising available landmarks. As a learning strategy, the authors adopted a mnemonic technique that allowed the patient to associate the street name with its location on the map. By applying this strategy to a limited range of streets in a familiar part of town, the patient was able to learn permanently the names and locations of the streets on the map.

Unfortunately, Davis and Coltheart's patient failed to generalise this strategy to new path learning and kept on using a landmark-based strategy even though it was ineffective.

Brunsdon and colleagues (2007) describe the assessment and treatment of a six-and-a-half-year-old child affected by topographical disorientation associated with visual agnosia and severe impairment in general spatial learning and memory due to an extensive intraventricular haemorrhage that occurred eight days after birth. Their treatment focused on improving the patient's topographical orientation in the school environment. First, they trained her to recognise major school buildings and landmarks and then to find commonly used routes in the area of the school. On one hand, this study has the merit of reporting a case of assessment and treatment of a childhood topographical disorientation never described until now; on the other hand, however, the patient does not show a selective topographical deficit as she suffers from a mild visual agnosia and more general spatial learning and memory deficits. Moreover, such a small child has very little autonomy in everyday life movements. For this reason, the authors limited the treatment to the school environment and taught the child's parents and school personnel how to apply a management strategy in the future. This type of treatment programme does not allow evaluation of real improvement in navigation ability and relies on the caregivers to intervene when the child is in difficulty.

The rehabilitation treatment reported in this paper is a case of topographical disorientation (Iaria, Incoccia, Piccardi, Nico, & Sabatini, 2005) that differs from others in the literature for several reasons. Selective topographical deficits are usually described in patients who have lost the ability to orient themselves in the environment as a consequence of acquired brain damage. In other words, they became impaired after their navigational skill was completely developed. However, due to congenital brain malformation our patient never developed orientation skills. One interesting aspect of this case is that the patient's selective impairment was confined to the spatial domain in spite of widespread brain malformation bilaterally involving the parietal, temporal and occipital cortex. Because MGC had not had an accident, one wonders why normal development of navigational skills was blocked rather than other cognitive functions. We hypothesised that the re-organisation of MGC's brain favoured more basic cognitive functions. In fact, in spite of the morphology of her posterior brain, MGC did not develop any visual field defects and did not show any agnosic or apractic deficits, indicating spared functional development of the primary and associative visual cortex. This suggests that brain functions which develop at a later stage of cognitive development suffered from the priority given to primary and earlier cognitive functions. Furthermore, as MGC's clinical history started very early in her life she never had the chance to practise spatial competence, as healthy children do, making it impossible for her to develop a complete cognitive system to orient and navigate within the environment.

For these reasons, the rehabilitation treatment of her topographical disorder was not aimed at re-educating a complex cognitive function compromised by acquired brain damage or at re-acquiring lost abilities. Rather, the goal was to make it possible for the patient to acquire navigational skills she had never developed by teaching her to use spatial language and verbal strategies. The final aim was to make it possible for MGC to navigate not only in familiar environments, but also in previously unknown environments.

CASE HISTORY

MGC is a 20-year-old, right-handed (Salmaso & Longoni, 1985) woman. When she was 21 days old, she underwent insertion of a ventricular-peritoneal shunt because of congenital hydrocephalus. At six months of age, MGC, who was suffering from meningitis, underwent brain surgery to monitor the shunt draining system. Afterwards, she showed unilateral partial motor seizures that were pharmacologically treated with anticonvulsant medication for three years. During the course of her life, MGC has undergone brain surgery to monitor the shunt on several occasions (last surgery in July 2001).

MGC's motor development was within the normal range: she obtained trunk control at the age of 6 months and learned to walk before 2 years of age. Her language development was normal and she apparently reached normal cognitive levels. In fact, MGC's school attendance was regular and she completed high school successfully.

Despite her normal cognitive development, MGC reported that she had never been able to orient herself in the environment. Although she is able to find her way around the six-room apartment she has lived in since she was born, MGC never leaves home alone because she always gets lost. She also gets lost if she loses sight of her mother in the grocery store. At the end of high school, following a long period of training with her father, MGC learned two short routes: starting from the main entrance of her school, she is able to walk straight ahead to the end of the main street, where she turns right to reach her father's shop, 200 metres away, or turns left to reach her grandmother's house, at about the same distance (see Figure 1).

In December 2002, she was referred to our centre for evaluation of her topographical disorder. We submitted MGC to a neuroradiological examination, a complete neuropsychological assessment and an evaluation of the specific abilities involved in topographical orientation. We obtained the patient's informed consent, as required by the local ethics committee.

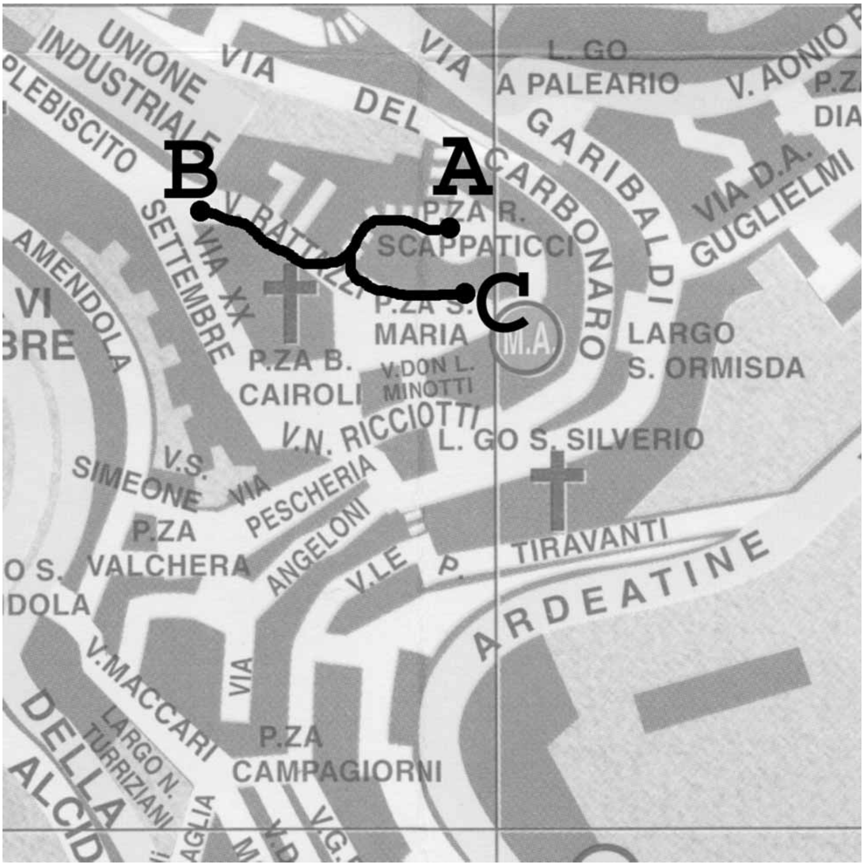

Figure 1. The black lines indicate the only two paths MGC had learned in her life: Starting from her high school (A) to her father's shop (B; approx 250 m) or to her grandmother's home (C; approx 200 m).

Neuroradiological examination

MRI scans showed an association between malformations of the posterior fossa and supra-tentorial abnormalities (see Iaria et al., 2005 for further details). The malformations of the posterior fossa included a vermian-cerebellar hypoplasia with associated large retro-cerebellar cerebral-spinal fluid collection and elevation of the tentorium and torcular, as well as dysmorphia of the fourth ventricle. The supra-tentorial malformations included hypoplasia of the rostrum, the posterior part of the corpus and the splenium of the corpus callosum, thickening of the white and grey matter of the right medial temporal and occipital lobes, and a cortical dysplasia (polymicrogyria) of the left medial temporal-parietal-occipital lobes. In the temporal lobe, a craniotomic foramen was present due to placement of the ventriculoperitoneal shunt; the ventricular catheter entered into the right trigone and terminated in the left trigone. Both posterior horns of the lateral ventricles appeared dysmorphic, with reduced size and irregular wall contours. The loss of the deep white matter surrounding the posterior horns may be the result of periventricular leukomalacia. Overall, the MRI study suggests complex anomalies of the cerebellum, the cerebral commissural system and the cortical organisation of both the temporal and the occipital lobes.

Neuropsychological and navigational skills assessment

MGC was submitted to a complete neuropsychological assessment of general intelligence, attention, memory and visuo-spatial perception (for a detailed description of assessment see Iaria et al., 2005). On all tests for which there is no published normative data, the patient's performances were compared to those of a control group matched for sex, age, and education (see Iaria et al., 2005).

The patient had normal verbal fluency and comprehension. On the Wechsler Adult Intelligence Scale–Revised (WAIS-R; Italian version, Orsini, & Laicardi, 1997) she obtained a Verbal IQ of 94 and a Performance IQ of 78 (total IQ = 86). MGC did not show any signs of apraxia (ideo-motor, ideational or constructional), unilateral neglect, visual imagery defect or agnosia for faces, colours or objects. However, she performed below the cutoff on Benton's Visual Retention Test (Benton, Levin, & Van Allen, 1974), the Judgment of Line Orientation Test (Benton, Varney, & Harmsher, 1978), the Cube Analysis subtest of the Visual Object and Space Perception (VOSP) battery (Warrington & James, 1991), the Corsi Supraspan Block Test (Spinnler & Tognoni, 1987) and delayed recall of the Rey-Osterrieth Complex Figure (Osterrieth, 1944) (see Table 1, pre-treatment). These tests were re-administered after the rehabilitation treatment.

MGC underwent a battery of tests to assess her navigational skills (see Table 2, pre-treatment). These included real navigation tasks in both experimental and ecological environments to document the well-established deficit in navigation reported by both the patient and her parents.

Following the guidelines on the evaluation of the effectiveness of cognitive rehabilitation training (Cicerone et al., 2000), testing was devised so as to allow retesting at the end of the treatment and follow-up with tasks that would be effective in measuring the efficacy of the treatment for acquiring navigational competencies and their generalisation to everyday life. We decided not to apply a double-baseline assessment as it was unnecessary to exclude spontaneous recovery or after-effects of previous trainings in a patient affected by a congenital disorder with unchanging severity.

TABLE 1
Neuropsychological assessment pre- and post-treatment

Test	*Pre-treatment*	*Post-treatment*	*Cutoff*
General intelligence			
Wechsler Adult Intelligence Scale (Wechsler, 1955)			
Verbal IQ	94		< 75
Performance IQ	78		< 75
Full scale IQ	86		< 75
Memory			
Verbal memory			
Rey's 15 Words Learning Task (Carlesimo et al., 1996)			
Immediate recall	29.9		28.53
15 min. delayed recall	5.2		4.69
Digit Span (Orsini et al., 1987)			
Forward	6		3.75
Backward	4		
Short Story Immediate recall (Novelli et al., 1986)	11.5		10
Spatial memory			
Corsi Block Test (Spinnler & Tognoni, 1987)	3.5		3.5
Corsi Block Test Supra-Span (Spinnler & Tognoni, 1987)	**0.92***	**3.51***	5.5
Rey's Figure A (delayed recall) (Osterrieth, 1944)	Refused*	**12 < 10° cent***	15 = 10°cent
Visual memory			
Benton's Visual Retention Test (Benton et al., 1974)	6/10		5/10
Attention			
Visual search (Spinnler & Tognoni, 1987)	52/60		39.25
Visual-perceptual abilities			
VOSP (Warrington & James, 1991)			
Object perception			
Screening Test	20/20		15/20
Incomplete Letters	18/20		17/20
Silhouettes	17/30		16/30
Object Decision	19/20		15/20
Progressive Silhouettes	10/20		14/20
Space perception			
Dot Counting	10/10		8/10
Position Discrimination	18/20		18/20
Number Location	8/10		7/10
Cube Analysis	**4/10***	**4/10***	6/10
Judgement of Line Orientation (Benton et al., 1978)	**14/30***	**17/30***	19/30
Street's Completion Test (Spinnler & Tognoni, 1987)	8/14		2/14

(*Continued*)

TABLE 1
Continued

Test	*Pre-treatment*	*Post-treatment*	*Cutoff*
Unfamiliar Perspectives	11/16		8.5/16
Rey's Figure A (copy) (Osterrieth, 1944)	33/36		29/36
Neglect battery (Pizzamiglio, et al., 1990)			
Line Cancellation	21/21		21/21
Letter Cancellation	104/104		100/104
Wundt-Jastrow Area Illusion Test	20/20		20/20
Reading Test	6/6		6/6
Line Bisection Test	+		
Imagery abilities			
O'clock Test (Grossi et al., 1989)	27/32		
Shape Recognition (Grossi, 1991)	9/10		8.45/10

*performance below the cutoff.

The neuropsychological evaluation included the assessment of specific cognitive abilities relevant for navigation.

1. Mental rotation (Mental Rotation Test; Trojano et al., 2004), mental representation of a familiar environment (Map Drawing), body displacements according to a paper map (Road Map Test; Money, Alexander, & Walker, 1965) and processing of vestibular information in a path estimation task (Distance Replication Test; Pizzamiglio, Iaria, Berthoz, Galati, & Guariglia, 2003);
2. Investigation of specific navigational processes in an experimental environment. These included the ability to use idiothetic or allothetic information to reach a target location (Place Learning Test in Experimental Environment, Guariglia, Piccardi, Iaria, Nico, & Pizzamiglio, 2005; Nico et al., 2008) and the ability to translate a schematic representation (a map) into locomotion (Semmes Test; Semmes, Weinstein, Ghent, & Teuber, 1955);
3. Evaluation of navigational skills in ecological environments by testing the ability to use a map (Map-Based Way Finding), to learn a path using different strategies (Route-Based Way Finding, Landmark-Based Way Finding) and to recognise salient elements available in the environment and useful for orientation (Landmark Identification).

Next we briefly describe the procedure, requirements and MGC's performance on each task included in the navigational battery.

MGC's performance was impaired on a mental rotation test (Trojano et al., 2004) when rotational disparity between targets and items exceeded 90°.

TABLE 2
Navigational skills assessment. Tasks are grouped according to relative cognitive abilities

Test	*Pre-treatment*	*Post-treatment*	*Cutoff (SD)*
Mental Rotation (Grossi, 1991)	**6/10***	**8/10***	9.07 (1.72)
Mental representation			
Map Drawing	−	+/− (improved performance with a survey perspective)	
Road Map Test (Money et al., 1965)	**14/32***	**20/32***	22/32
Distance Replication (Pizzamiglio et al., 2003)	*M (SD)*		*Control M (SD)*
Leftward	**1.51 (0.40)***		2.83 (0.07)
Forward	**2.45 (0.24)***		3.02 (27.7)
Rightward	**0.75 (0.92)***		2.97 (0.33)
Place Learning Test in experimental environment (Guariglia et al., 2005)			
No landmark	*M (SD)*		*Control M (SD)*
Searching (seconds)	**80***		46.3 (31.3)
Immediate recall (seconds)	**131.8 (101.1)***		48 (27.7)
Delayed recall (seconds)	**28***		26.3 (4.3)
With landmark	*M (SD)*		*Control M (SD)*
Searching (seconds)	**428***		24
Immediate recall (seconds)	**224 (105.6)***		25.8 (9.9)
Delayed recall (seconds)	**68***		17.3 (6.2)
Semmes Test (Semmes et al., 1955)	**0/5***	4/5	
Way Finding in ecological environment			
Map-Based	−	+	
Route-Based	−	+	
Landmark-Based	−	+	
Landmark Identification	+	+	

− impaired performance. *performance below the cutoff.

She was unable to draw a map of her home from memory. Although she correctly reported the number of rooms in the apartment, she produced a map that was distorted and that contained errors of scale and of spatial relations among elements (see Figure 2a).

In the Road Map Test (Money et al., 1965), the patient was required to imagine she was moving using an A4-sized paper map. As she followed the traced pathway, she had to report verbally whether she would go left or

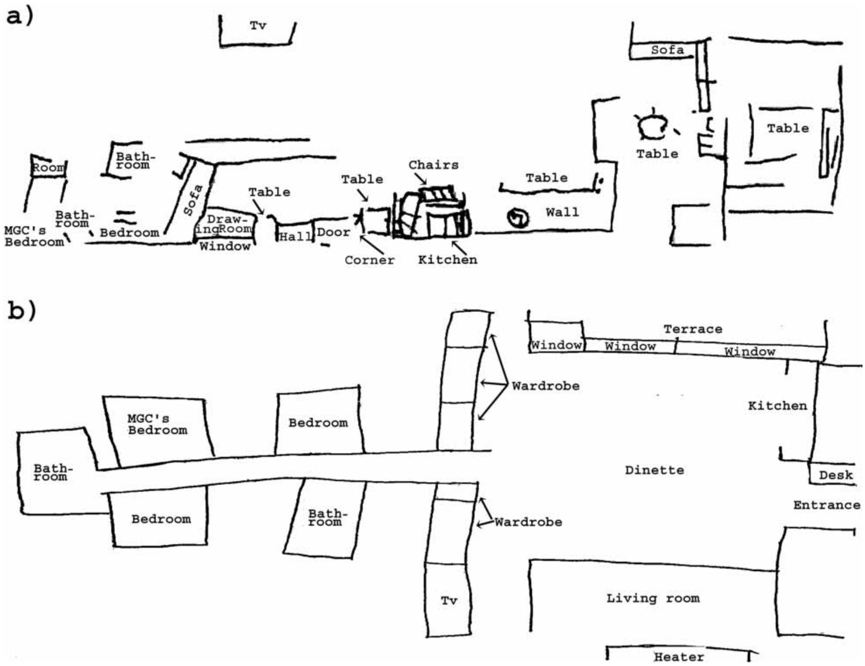

Figure 2. MGC's map-drawing of her home: (a) pre-treatment; (b) post-treatment. In the pre-treatment drawing map (a), rooms and furniture were drawn based on verbal recall of items, without regard to their spatial organisation. On the contrary, in the post-treatment map (b), although her graphic representation was still defective, the actual spatial organisation of rooms and furniture was respected.

right at each change of direction on the map. MGC judged only 14 out of 32 turns (cutoff 22) correctly; thus, her performance was clearly impaired.

The processing of vestibular and somatosensory information, which subserves the ability to evaluate distances and path lengths, was assessed using the Distance Replication Test (Pizzamiglio et al., 2003). MGC was asked to replicate actively a linear displacement (three metres) she had passively undergone to the left, the right or forward. The test was performed in an empty room devoid of visual cues. MGC's responses were shorter than those of controls in all directions.

The Place Learning Test in an experimental environment (Guariglia et al., 2005; Nico et al., 2008) was used to assess three basic navigational systems: path integration, re-orientation and landmark-based navigation (Wang & Spelke, 2002). These systems are based on the processing of idiothetic, geometric and visuo-spatial environmental information, respectively. The task was performed in an empty, 5 × 6 m room in which no visual cues were available. The subject was asked to find a hidden location which, when found, was indicated by an acoustic stimulus (searching condition). As soon as the subject reached the hidden location she was blindfolded, disoriented, moved back to the centre of the room and asked to find the same hidden location by following the shortest path (immediate recall condition, six trials). Thirty minutes later, the subject performed one recall trial (delayed recall condition). In a different session, two landmarks (a lamp and a clothes hook) were present in the room. This time the hidden target location was different. The same tasks were repeated.

MGC performed the tasks more slowly than the control subjects in both conditions, i.e., with and without landmarks. Moreover, unlike the control subjects, she did not take advantage of the landmarks to solve the task. In fact, in this condition the control subjects took half the time they had taken in the no-landmarks condition; MGC, on the contrary, took twice as long.

In the Semmes Test (Semmes et al., 1955), MGC was given a schematic drawing of a 3 × 3-point grid and she had to translate the drawn lines on the map into locomotion by walking the designated path on a 3 × 3-point grid laid out on the floor (3 m × 3 m). In this classic test, which assesses the ability to use a map for real navigation, MGC was completely unable to reproduce any of the five paths.

In the Map-Based Way Finding Test, the patient was given a city map on which two spots were indicated: her actual position and the goal. The patient had to indicate the shortest path (about 200 m) on the map and then follow the path to reach the final location. She failed on this task by getting lost as soon as a turn on the map indicated a change of direction. For approximately two hours, she made repeated attempts without ever finding the goal.

A Route-Based Way Finding task was administered in which MGC had to replicate a route she had followed before. The experimenter led the subject along the route (about 200 m long; three left and three right turns) to the final location without giving her any verbal information. Then she was guided back to the starting point by means of a shortcut and, immediately after, was asked to reach the final location by following the same path. The test was carried out in two different places and two different sessions: near the hospital (route A) and downtown (route B, see Figure 3a). The whole procedure was repeated three times for each route. MGC never succeeded in reaching her destination; she felt disoriented at the beginning of each route and randomly turned right or left.

A Landmark-Based version of the previous test was developed, as MGC was completely unable to reproduce even the simplest route. The procedure was identical to the previous one except that, at each turn, the experimenter indicated a specific landmark (selected from many available on the street) useful for orientation and asked the patient to examine it carefully and then describe it verbally. The task was performed downtown on a new route (Route C), which was similar to Route B used in the previous task. Again, MGC did not follow the route and never reached the target location

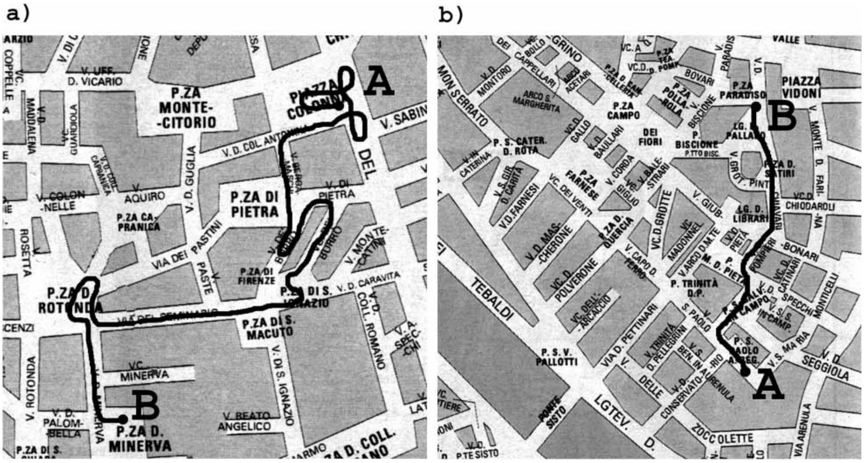

Figure 3. The figure shows examples of MGC's performances during the assessment of place learning in the ecological environments (a) before and (b) after treatment. The examples refer to the Map-Based Way Finding Task, which is the most difficult task on the ecological environment because it requires planning and following a route on a map from a starting point (A) to a final destination (B)

because she got lost at each turn. Even though she promptly recognised the relevant landmark at each turn, she was unable to derive any directional information from it.

The patient's ability to recognise 36 pictures, including landmarks and views previously seen while following Routes A, B and C and interspersed with 24 distractor pictures of a different area of Rome the patient had never been to, was evaluated one week later. She correctly recognised 81% of the familiar items and 54% of the unfamiliar ones. This result, which partially contrasted with her above reported ability to recognise landmarks while following routes, could be accounted for by the similarity (see Figure 4) between distractors and targets on one hand and the distance from learning to recognition on the other.

In summary, the neuropsychological assessment showed no general cognitive impairments. She performed within the normal range on all tasks except visuo-spatial memory, mental rotation and line orientation. These deficits affected her ability to use a map and to understand the correspondence between the real environment and its graphical representation on the map.

Some of the basic abilities useful for orientation were also spared (e.g., complex visuo-perceptual skills). Even though her proprioceptive, vestibular, visual and perceptual abilities were spared, she showed a deficit in integrating all this information to build up a mental representation of the environment.

Figure 4. Example of landmark picture and its distractor.

The peculiarity of our patient was that she had not acquired any experience in topographical orientation and she lacked all the navigational competencies individuals normally develop during their lifetime.

With reference to Aguirre and D'Esposito's taxonomy, we identified the deficit showed by MGC as a mixed disorder. Indeed, MGC was able to recognise landmarks in the environment, but she failed to code their position relative to herself and to derive directional information from them. Moreover, she seemed unable to learn pathways in novel environments. Thus, we posited that her disorientation was threefold: egocentric, heading and anterograde. However, based on De Renzi's model we classified our patient as having the topographical amnesic disorder and ruled out the topographical agnosic component.

In any case, it is difficult to fit a developmental disorder in a taxonomy describing acquired deficits as, for example, in the case of developmental language disorders (dyslexia or dysphasia) and acquired language deficits.

Treatment

The final aim of the treatment was to bring MGC to the point of developing a navigational strategy she could use by herself so that, besides being able to move in familiar environments, she would also be able to reach new, unfamiliar places by following paths she had never before taken.

As it is currently accepted that deficits can never be completely recovered (Parente & Stapleton, 1997), our aim was not to restore MGC's topographical abilities but, rather, to familiarise her with alternative strategies (e.g., language-based navigation).

To overcome her poor visuo-spatial abilities in mental rotation, spatial memory and visuo-spatial reasoning, which led to imprecise and therefore ineffective use of spatial language, in the first 10 sessions we trained some basic visuo-spatial abilities essential for correctly exploring, memorising and recognising environmental features.

In teaching MGC to navigate by means of a language-based strategy, she was first trained to use verbal instructions to follow a path and then trained to create the instructions necessary for describing paths in novel environments. Throughout the entire treatment, MGC was continuously encouraged to use any of her newly acquired skills in daily life to increase her autonomy.

The next two sections describe the exercises used to train MGC's visuo-spatial abilities and language-based navigation.

Training of visuo-spatial abilities. As MGC lacked practice, we trained her to observe carefully different environments; this preliminary work was aimed at teaching the patient to look around and to distinguish the available landmarks. In particular, we proposed tasks in which the patient had to recognise differences in pairs of pictures representing two similar environments (20 items) or two mirror-image versions of the same environment (20 items).

As MGC's spatial representation ability was poor, we trained her to describe familiar places (home, hospital area, etc.) and to locate various environmental elements. For example, she was taken into a room and asked to indicate the locations of the elements that were out of sight (e.g., entrance, staircase, telephone, toilet, etc.). Prior to this step, it was necessary to teach her to define spatial relations among elements (e.g., "The window is in front of the door", "The table is on the left wall of the room", etc.).

To improve her mental rotation skills, the patient was trained (first with real objects and then with abstract pictures) to observe objects and then to imagine their position if they were rotated to different angles (45°, 90°, 135°, 180°, 270°). Exercises involving body rotation with respect to the environment were also proposed to help her understand the invariance in the positions of the environmental elements in spite of body displacements. In this exercise, MGC was asked to point to the positions of various elements (out of sight) before and after her body was rotated.

A large part of the work consisted of learning to read, interpret, and subsequently draw internal and external environmental maps. We had to take into account that MGC had never learned to use a map and that she was initially unable to grasp the idea of the possibility of representing a site from a survey perspective.

Examples of the tasks used in the rehabilitation treatment are reported in the Appendix.

By the end of this step, MGC had learned to observe and describe in detail environments and their elements. In fact, after initial difficulty on the picture differences recognition tasks of similar environments she correctly performed 18 out of 20 items; in the mirror version, she correctly recognised 20 out of 20 pictures.

The patient also learned to describe the spatial relations among the landmarks in familiar environments (e.g., "The coffee shop is to the right of the Day Hospital entrance and in front of the Infant Rehabilitation Unit"). It was evident, however, that despite her improved ability to describe environmental features, the patient was still unable to represent mentally all kinds of places, including those she was most familiar with (e.g., her home). In fact, she still failed in drawing maps and in describing the spatial relationships between objects in environments when they were out of sight (see Figure 5).

MGC was able to draw small maps, but only when she was prompted by a verbal description. Therefore, she was unable to imagine an environment and was able to draw it only if the examiner tested her knowledge by asking some questions (see the Appendix for details). Although she demonstrated good

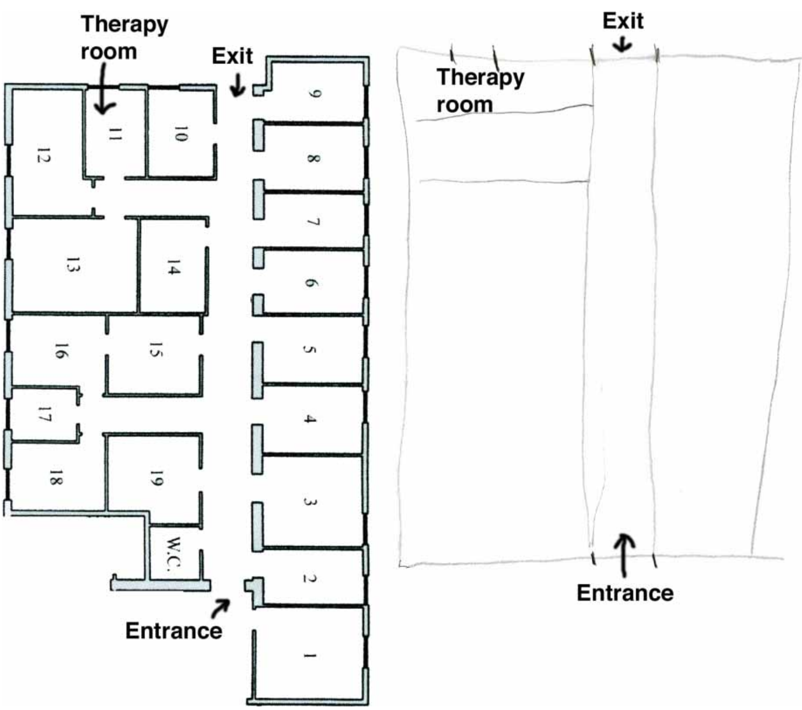

Figure 5. Map of the Neuropsychology Unit and MGC's drawing from memory.

semantic knowledge of the environment, her performance was very poor and inaccurate due to alterations in proportions and spatial relationships.

The mental rotation exercises only partially improved her deficit. MGC was only able to rotate mentally objects located in front of her; she could not make complex rotations such as those required for imagining environmental changes after a body displacement. In the object rotation task, MGC correctly performed only 90° and 180° rotations, and failed on 45°, 135° and 270° rotations (the exercise was composed of 10 items for each rotation). She performed 20 out of 50 items without error. In the body rotation task (60 items), she was only able to solve items involving 180° rotations (20 out of 60) and failed on 90° and 45° body rotations.

In spite of the map training, MGC was still unable to create cognitive maps of places or to use maps to follow a path; indeed, she could not even follow five very easy paths inside the hospital using the map (0 out of 5).

Training of language-based navigation. The exercises proposed in the second step were aimed at functionally treating MCG's topographical disability.

First of all, MGC was taught to follow written instructions, to walk short distances inside the hospital, and to look around and observe the elements she encountered along the way that could be used during navigation.

Subsequently, she was requested to write descriptions of the paths inside the hospital and to identify landmarks and directions. The patient was accompanied along the selected route and she gradually wrote out the instructions.

To verify the efficacy and accuracy of the descriptions, in the next session she was asked to cover the same route using her own instructions. At this time, the therapist pointed out her errors and MGC corrected them.

As MGC mastered this technique, a large part of the work involved using maps of different areas of the city and trying to teach her a procedure to adopt for moving in unfamiliar environments. At the beginning, she had to localise specific starting and ending points on the map and she had to choose the best (or shortest) way from one to the other; then, she had to write down the verbal instructions for covering that distance. We had to help the patient write the information clearly and concisely. Very often her instructions were redundant or she failed to specify a landmark when she changed direction. For example, MGC wrote that she should turn left at the first opportunity. However, she failed to consider that the street she saw on the left side of the map might have a different location in the real environment and, therefore, had to be more clearly specified in the written description of the path (i.e., by indicating the name of the street). After several sessions of these exercises, she began to follow the described routes in the real environment.

In the first session outside the hospital, we had to deal with the patient's total inexperience of moving around by herself (e.g., we had to teach her where street names are usually located and suggested that she move to the right-hand side of the street when she had to find a street on the right).

During the early sessions, MGC frequently stopped or hesitated due to anxiety and had to be urged to continue. The therapists who accompanied the patient were trained to intervene as little as possible in her choices and just to encourage her to follow the instructions. They explained that even errors were useful because they prepared her for correct performances in the future.

Sometimes MGC had to face unexpected situations, e.g., work in progress that blocked the street she had to cross so that she was forced to find alternative solutions, such as looking at the map again or asking someone for directions. It was especially difficult to convince the patient to use this last solution, because she felt she was being observed and would be considered inadequate by other people.

After MGC became more confident and more self-reliant, we invited her mother to participate in the sessions. Our aim was twofold: to demonstrate MCG's potential to her mother and to teach the latter the procedure for describing and following the paths. We had to explain that she should not guide her daughter's steps, but should just assist and reassure her. From then on, thanks to the active participation of her mother, MGC began practising in her own city.

As the treatment proceeded, MGC not only showed improved navigational abilities but also a dramatic change in attitude. Although in the beginning she found moving in unfamiliar environments difficult and distressing, at the end she became enthusiastic about the task and happily recognised her new skill.

Final results

At the end of the treatment, the patient was submitted to all the tests on which her performance had been deficient at the first evaluation (see Tables 1 and 2, post-treatment).

Although her scores remained slightly below the cutoff on some tasks, her performances clearly improved in quality. For example, in the Road Map Test the patient made fewer errors in left/right discrimination, as shown by the increase in correct responses (see Table 1 post-treatment). Moreover, her ability to represent and reproduce her home map had changed considerably: Her performance was still schematic and very vague, but she was now able to create a mental image of the environment (see Figure 2b).

Such improvements in basic abilities allowed the patient to move in more ecological situations. Before treatment she was totally unable to use even the easiest map on the Semmes Test, but after treatment she correctly performed four pathways out of five. The most consistent results were observed in tasks measuring the patient's ability to use navigational skills autonomously in the real environment. In the final assessment, the patient was once again submitted to the ecological tasks, but using different environments and paths to those used in the first test and those trained during the treatment.

In the Way Finding Map-Based task, MGC was now able not only to choose the best path on the map, connecting the starting point with the final destination, but also to follow it without hesitation or errors (see Figure 3b). In the Way Finding Route-Based task a longer and more complex pathway was proposed, and MGC showed no difficulty in covering the distance she previously saw. It should be noted that during this task the patient spontaneously used landmarks and indicated them to the examiner as she walked. MGC's performance was similar when she was required to change direction and follow a previously covered distance backwards. As the patient showed good ability in spontaneously using landmarks, the Way Finding Landmark-Based task was not administered again.

The results showed great improvements on ecological tasks. Not only had the patient developed specific navigational skills, but her behaviour had changed. During her performances, MGC showed self-confidence. She now walked ahead of the examiner and looked around and observed the environment; prior to the treatment, she had passively followed the therapist.

Follow-up

A follow-up was performed approximately one year after the end of the treatment. At that time, a structured interview was also held to assess whether MGC was making autonomous use of the navigational competencies she had learned and to determine the frequency with which she went around by herself and which kind of autonomous navigation she usually adopted.

During the interview, the patient claimed that she had begun moving around her city. She took a bus whenever she needed to buy something or just wanted to go shopping downtown. With her improved navigation skills, MGC felt more confident in her abilities and was even attempting to take a computer course.

We again submitted the patient to three pathways in the real environment that were different from those she had previously travelled. In the first trial, she had to choose the best way to go from the starting point to the final destination on a city map and she had to write down the verbal instructions for covering that distance (see Figure 6). She then had to follow her own instructions to reach the final location. In the second and third tasks, respectively, we asked MGC to replicate a route she had previously travelled and then to go backwards. On all three pathways her result was the same as that reached the year before, showing she was able to follow a new route using her own

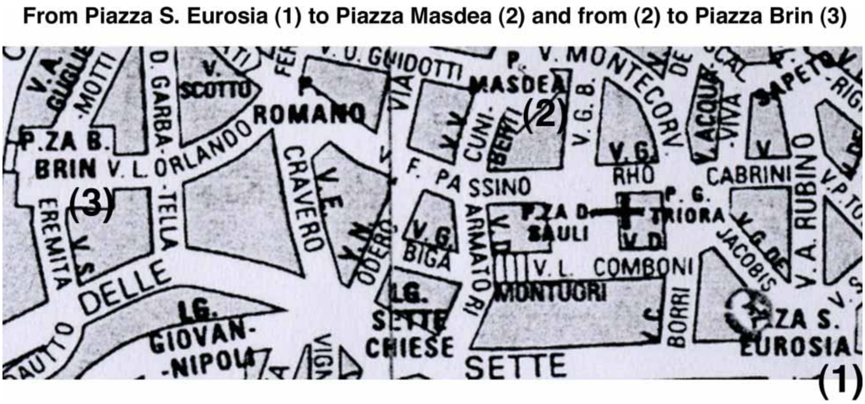

Figure 6. This figure depicts the map with the path that MGC had to: (a) verbally describe, (b) write instructions about following, and (c) follow. The written instructions followed by MGC were: "*From Piazza Sant'Eurosia (1) take Via De Jacobis, which arrives in Piazza Triora; from there go straight ahead to Via Montecorvo, which leads to Piazza Masdea (2). From there take Via Guidotti to Via Orlando Ferrati, turn left and go toward Piazza Brin (3).*"

instructions and to correctly cover the distances we had proposed both forwards and backwards.

MGC still feels anxious, vulnerable and insecure. However, she is able to overcome these feelings and has become much more autonomous in her movements.

DISCUSSION

In this paper, we describe the treatment of topographical disorientation in a young woman with a congenital malformation who never developed the ability to navigate by herself. Her disorder was characterised by egocentric and heading disorientation and by inability to use a map and to understand the correspondences between real environments and their graphic representations. To plan a suitable rehabilitation intervention for the patient, we had to assess the cognitive skills involved in orienting processes and to investigate the functional abilities she was using to navigate in the environment.

Overall, the assessment revealed that the patient's deficit was a consequence of the alteration of representational skills of space and topographical coordinates that are necessary for navigation and also of the lack of development of complex processes integrating all of the components involved in space orientation and navigation.

The treatment had different aspects: One focused specifically on several defective visuo-spatial abilities necessary for navigation, in which the patient showed only limited improvement (i.e., mental rotation). The other aspects were more functional and ecological: the patient was trained to develop compensatory strategies and to process language-based navigation, which gradually allowed her to become independent.

Our successful results in this case, which involved very complex and multi-component cognitive functions, confirm the importance of making a very detailed assessment of the patient's competencies and behaviour in ecological situations. Using this kind of approach, it is possible to evaluate a patient's actual abilities and plan the most effective treatment.

Our approach was effective because it allowed MGC to learn navigational skills that permitted her to move by herself even in previously unknown environments. Also, not only did MGC's acquired abilities remain unchanged after the end of the treatment, but also she showed a trend towards continuous, slight improvement because daily use of autonomous navigation is an effective exercise.

It is noteworthy that the patient's defective visuo-spatial abilities (i.e., mental rotation, visuo-spatial memory), considered a basic component of the visuo-spatial competencies underlying navigation, underwent only minimal, ineffective changes following treatment. In fact, rather than reduce MGC's basic visuo-spatial deficits, the training allowed her to become aware of her difficulties and, therefore, to recognise and cope with them. MGC's newly acquired awareness allows her to avoid errors in daily life navigation. For example, as she is aware of her poor mental rotational abilities MGC provides herself with verbal descriptions of the appearance of landmarks and views from the opposite perspective so that when she returns along a new route she recognises them and does not get lost.

This study demonstrates, once again, that it is possible to obtain considerable improvement through appropriate assessment and treatment planning. More specifically, it is not only possible to rehabilitate lost abilities but also to encourage patients with congenital and seriously disabling cerebral damage to develop new skills.

REFERENCES

Aguirre, G. K., & D'Esposito, M. (1999). Topographical disorientation: A synthesis and taxonomy. *Brain*, *122*, 1613–1628.

Barrash, J. (1998). A historical review of topographical disorientation and its neuroanatomical correlates. *Journal of Clinical and Experimental Neuropsychology*, *20*(6), 807–827.

Benton, A. L., Levin, H. S., & Van Allen, M. W. (1974). Geographic orientation in patients with unilateral cerebral disease. *Neuropsychologia*, *12*, 183–191.

Benton, A. L., Varney, N. S., & Hamsher, K., de S. (1978) Visuospatial judgment: A clinical test. *Archives of Neurology*, *35*, 364–367.

Brunsdon, R., Nickels, L., Coltheart, M., & Joy, P. (2007). Assessment and treatment of childhood topographical disorientation: A case study. *Neuropsychological Rehabilitation*, *17*(1), 53–94.

Carlesimo, G. A., Caltagirone, C., & Gainotti, G. (1996). The mental deterioration battery: Normative data, diagnostic reliability and qualitative analyses of cognitive impairment. *European Neurology*, *36*, 378–384.

Cicerone, K. D., Dahlberg, C., Kalmar, K., Langenbahn, D. M., Malec, J. F., Bergquist, T. F., Felicetti, T., Giacino, J. T., Preston Harley, J., Harrington, D. E., Herzog, J., Kneipp, S., Laatsch, L., & Morse, P. A. (2000). Evidence-based cognitive rehabilitation: Recommendations for clinical practice. *Archives of Physical Medicine Rehabilitation*, *81*, 1596–1615.

Davis, S. J. C., & Coltheart, M. (1999). Rehabilitation of topographical disorientation: An experimental single case study. *Neuropsychological Rehabilitation*, *9*(1), 1–30.

De Renzi, E. (1982). *Disorders of Space Exploration and Cognition*. Chichester, UK: Wiley.

Farrell, M. J. (1996). Topographical disorientation. *Neurocase*, *2*, 509–520.

Grossi, D. (1991). *La riabilitazione dei disturbi della cognizione spaziale*. Masson, Milano.

Grossi, D., Modafferi, A., Pelosi, L., & Trojano, L. (1989). On the different roles of the cerebral hemispheres in mental imagery: The "o'clock test" in two clinical cases. *Brain and Cognition*, *10*, 18–27.

Guariglia, C., Piccardi, L., Iaria, G., Nico, D., & Pizzamiglio, L. (2005). Representational neglect and navigation in real space. *Neuropsychologia*, *43*(8), 1138–1143.

Habib, M., & Sirigu, A. (1987). Pure topographical disorientation: A definition and anatomical basis. *Cortex*, *23*, 73–85.

Iaria, G., Incoccia, C., Piccardi, L., Nico, D., Sabatini, U., & Guariglia, C. (2005). Lack of orientation due to a congenital brain malformation: A case study. *Neurocase 11*, 463–474.

Money, J., Alexander, D., & Walker, H. T. (1965). *A Standardized Road Map Test of Direction Sense*. Baltimore: The Johns Hopkins Press.

Nico, D., Piccardi, L., Iaria, G., Bianchini, F., Zompanti, L., & Guariglia, C. (2008). Landmark based navigation in brain-damaged patients with neglect. *Neuropsychologia*, *46*, 1898–1907.

Novelli, G., Papagno, C., Capitani, E., Laiacona, M., Vallar, G., & Cappa, S. F. (1986). Tre test clinici di memoria verbale a lungo termine. Taratura su soggetti normali. *Archivio di Psicologia, Neurologia e Psichiatria*, *47*, 278–296.

Orsini, A., Grossi, D., Capitani, E., Laiacona, M., Papagno, C., & Vallar, G. (1987). Verbal and spatial immediate memory span: Normative data from 1355 adults and 1112 children. *Italian Journal of Neurological Science*, *8*, 539–548.

Orsini, A., & Laicardi, C. (1997). *WAIS–R Contributo alla taratura Italiana*. Firenze: O.S. Organizzazioni Speciali.

Osterrieth, P. (1944). Le test de copie d'une figure complexe. *Les Archieves de Psychologie*, *31*, 206–356.

Pallis, C. A. (1955). Impaired identification of faces and places with agnosia for colours. *Journal of Neurology, Neurosurgery and Psychiatry*, *18*, 218–224.

Parente, R., & Stapleton, M. (1997). History and systems of cognitive rehabilitation. *NeuroRehabilitation*, *8*, 3–11.

Pizzamiglio, L., Antonucci, G., Guariglia, C., Judica, A., Montenero, P., Razzano, C., & Zoccolotti, P. (1990). *La rieducazione neurocognitiva dell'eminattenzione in pazienti con lesione cerebrale unilaterale.* Milano: Masson.

Pizzamiglio, L., Iaria, G., Berthoz, A., Galati, G., & Guariglia, C. (2003). Cortical modulation of whole body movements in brain damaged patients. *Journal of Clinical and Experimental Neuropsychology*, *25*, 769–782.

Salmaso, D., & Longoni, A. M. (1985). Problems in the assessment of hand preference. *Cortex*, *21*, 533–549.

Semmes, J., Weinstein, S., Ghent, L., & Teuber, H. L. (1955). Spatial orientation in man after cerebral injury: I. Analyses by locus of lesion. *Journal of Psychology*, *39*, 227–244.

Spinnler, H., & Tognoni, G. (1987). Standardizzazione e taratura italiana di test neuropsicologici. *Italian Journal of Neurological Science (Suppl 8)*, 1–120.

Stark, M., Coslett, H. B., & Saffran, E. M. (1996). Impairments of an egocentric map of locations: Implications for perception and action. *Cognitive Neuropsychology*, *13*, 481–523.

Takahashi, N., Kawamura, M., Shiota, J., Kasahata, N., & Hirayama, K. (1997). Pure topographic disorientation due to right retrosplenial lesion. *Neurology*, *49*, 464–469.

Taulbee, L. R., & Folsom, J. C. (1968). Reality orientation for geriatric patients. *Hospital and Community Psychiatry*, *17*, 133–135.

Trojano, L., Fragassi, N. A., Chiacchio, L., Izzo, O., Izzo, G., Di Cesare, G., Cristinzio, C., & Grossi, D. (2004). Relationships between constructional and visuospatial abilities in normal subjects and in focal brain-damaged patients. *Journal of Clinical and Experimental Neuropsychology*, *26*, 1103–1112.

Wang, R. F., & Spelke, E. (2002). Human spatial representation. *Trends in Cognitive Sciences*, *6*, 376–382.

Warrington, E. K., & James, M. (1991). *The Visual Object and Space Perception Battery.* Bury St Edmunds, UK: Thames Valley Test Company.

Wechsler, D. (1955). *Wechsler Adult Intelligence Scale.* New York: Psychological Corporation.

Manuscript received July 2007
Revised manuscript received May 2008
First published online September 2008

APPENDIX

Map reading

To help the patient understand the correspondence between the real environment and its representation on a map, she was taken to the top floor of the hospital and told to observe the spatial/topographical relationships among the different hospital buildings (Research Centre, Day Hospital, Neuropsychology Unit). Next she was asked to draw them on a sheet of paper, respecting their positions and proportions. She was then trained to read the actual floor plan of the hospital and to look for the main points of reference (e.g., entrance, emergency exit, staircases, lifts). Finally, she was asked to plan the route from the entrance to the emergency exit, describing each landmark she would encounter. She subsequently followed the route.

Map drawing

The following is an example of dialogue between MGC and the therapist as MGC drew the map. The therapist asked her to draw a map of the

Neuropsychology Unit, which she had previously observed and described very carefully.

MGC: I really don't know. . .. I can't do it.

Therapist: Okay, MG, what's the first thing you see when you enter the Neuropsychology Unit?

MGC: The entrance and then a corridor.

Therapist: Okay, draw them. . .and what's beyond the corridor?

MGC: Some rooms on the left and the right.

Therapist: Okay, go ahead. . .

MGC drew some rooms randomly along the corridor without regard to their proportions or spatial relationships.